

Focal Cortical Dysplasia with Dysplastic Neuroepithelial Tumor: Neuroimaging Features of Epilepsy's Hidden Pair

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Introduction

Accurate lesion characterization on magnetic resonance imaging (MRI) is essential for surgical planning in drug-refractory epilepsy (DRE) as this may impact seizure outcomes. Recognizing the presence of focal cortical dysplasia (FCD) alongside tumors is vital for ensuring optimal resection and minimizing recurrence.

Case

A 15-year-old male presented with DRE for 4 years, experiencing daily seizures. Semiologies included right-sided focal

motor seizures, behavioral arrest, and vertiginous auras with backward falls. Video electroencephalogram showed a left temporo-parietal focus.

MRI revealed a cystic lesion in the *left high parietal cortex* consistent with dysembryoplastic neuroepithelial tumor (DNET; ► **Fig. 1**). Additionally, a hyperintense T2 signal extended from the cortex to the ventricular wall, showing the characteristic “transmantle sign” of FCD (► **Fig. 2**). The patient underwent an electrocorticography-guided lesion resection. Histopathology showed dyslamination and hypoplasia surrounding the tumor with oligodendroglioma in a mucoid matrix, suggestive of FCD along with the DNET. Six months post-surgery, he was seizure-free, achieving Engel class IA outcome.

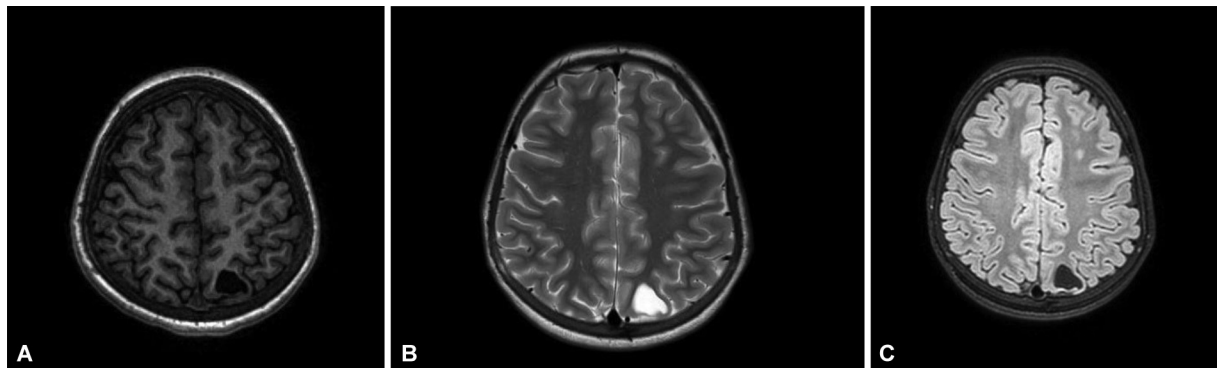


Fig. 1 MRI brain axial T1 sequence (A) demonstrating a cystic hypo-intense lesion. T2 sequence (B) demonstrating hyper-intense appearance of the DNET and FLAIR suppression (C). DNET, dysembryoplastic neuroepithelial tumor; FLAIR, fluid-attenuated inversion recovery; MRI, magnetic resonance imaging.

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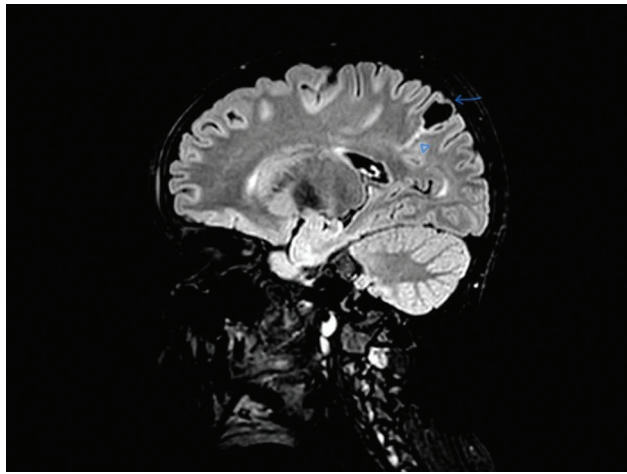


Fig. 2 Sagittal section of the brain with FLAIR-sequence demonstrating a FLAIR-suppressed oval cystic lesion (lobulated) in the *left parietal* region (*arrow*), with a hyper-intense signal (*arrowhead*) extending from its inferior boundary toward the lateral ventricle, consistent with the “transmantle sign” of FCD. FCD, focal cortical dysplasia; FLAIR, fluid-attenuated inversion recovery.

Discussion

FCD associated with tumors is classified as FCD IIIb. Reported incidence of FCD with DNET varies (5–80%) due to changing classifications.¹ DNETs appear as lobulated, T2 hyperintense lesions with a “bubbly” appearance and are typically hypointense on T1 images. They rarely show edema or enhancement and may mold the surrounding skull.

The “transmantle sign,” hallmark of FCD II, is linked to better postsurgical outcomes. FCD IIIb often coexists with lesions like DNET, the second most common tumor in these cases after gangliogliomas.² Genetic studies suggest a shared mammalian target of rapamycin (mTOR) pathway dysregulation, implicating abnormal cortical development and organization.

It is important to recognize both the lesions as the epileptogenic zone may extend beyond the visible lesion, necessitating wider resections to prevent recurrence. Prognosis is generally favorable, with up to 87% of patients achieving Engel IA outcomes at 5 years.³

Conflict of Interest

None declared.

Acknowledgments

None.

References

- 1 Takahashi A, Hong S-C, Seo DW, et al. Frequent association of cortical dysplasia in dysembryoplastic neuroepithelial tumor treated by epilepsy surgery. *Surgical Neurology* 2005;64 (Suppl 5):419–427
- 2 Cossu M, Fuschillo D, Brammerio M, et al. Epilepsy surgery of focal cortical dysplasia-associated tumors. *Epilepsia* 2013;54 (Suppl 9):115–122
- 3 Santos MV, de Oliveira RS, Machado HR. Approach to cortical dysplasia associated with glial and glioneuronal tumors (FCD type IIIb). *Childs Nerv Syst* 2014;30(11):1869–1874